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Short Report

Klippel–Trenaunay Syndrome Associated with Abdominal Aortic Aneurysm

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ABSTRACT

The Klippel–Trenaunay syndrome is a rare disorder characterised by well-described bony and vascular (venous and lymphatic) anomalies. Its association with arterial aneurysms has only been reported in a dozen cases, in particular, in cerebral, renal and popliteal arteries. We report the case of a 35-year-old male patient, who presented with an 85-mm aorto-iliac aneurysm primarily suspected to be mycotic, in addition to a typical single lower extremity arteriomegaly. The patient was successfully treated by means of an allograft. This is considered to be the first reported case of Klippel–Trenaunay syndrome, associated with an aortic aneurysm.

Introduction

The Klippel–Trenaunay syndrome (KTS) has many known clinical features and is usually associated with: bony hypertrophy of a single extremity, varicose veins and certain vascular anomalies, involving mostly venous and lymphatic malformations. Only eight cases of cerebral aneurysms,[1] two cases of renal[2] aneurysms and one case of popliteal aneurysm[3] have been reported. However, until the present no case of an aorto-iliac aneurysm had been reported.

Report

A 35-year-old, male patient was admitted to our hospital, on the basis of a suspected deep venous thrombosis (DVT) of the lower left limb. In his past history, we found only a moderate use of tobacco. At the time of admission, clinical examination revealed voluminous varicosities on the dorsal side of the left foot and on the leg, developed from the great saphenous vein, with vascular dermatitis, oedema and venous ulcers but without angioma. There was no enlargement of the lateral embryonic vein. The presence of a small saphenous vein thrombosis extending to the popliteal and superficial femoral vein was confirmed by the ultrasonography. No deep venous abnormality was noted. As the DVT was associated with hypoxia, a computed tomography angiography (CTA) was performed to look for a pulmonary embolism and revealed an 85-mm aneurysm of the entire abdominal aorta, including the left iliac axis (Fig. 1). Overall, the CTA revealed a unilateral left arteriomegaly (20 mm). As the patient was hyperthermic (42 °C), with the presence of a lower limb entry point (ulcer) and the presence of multi-susceptible *Staphylococcus aureus* in his blood cultures, a mycotic aneurysm was suspected. The operation was performed using a retroperitoneal approach. The aortic and iliac aneurysms were incised; many samples of the aortic wall were sent to pathology and microbiology departments. An aorto-bi-iliac allograft was interposed, anastomosed distally to the right common iliac artery and to the left external iliac artery. The aneurysm of the internal iliac artery was also incised. The internal iliac artery was not reimplanted but ligated in the bottom of the aneurysm. The postoperative follow-up was free of complications. All peroperative bacteriological samples were negative. The lower limb venous anomaly was explored by an angio-magnetic resonance imaging (angio-MRI) and an ultrasonograph. A purely capillary venous malformation was found. Then, the diagnosis of the KTS was suspected and confirmed on lower extremity X-ray images, which showed a unilateral bone hypertrophy (Fig. 2).

Discussion

Because of the hyperthermia, it was legitimated to consider first the possibility of bacterial colonisation over a degenerative aneurysm. In addition, the aneurysm was fusiform with some calcifications and many thrombi, which could suggest an atherosclerotic origin. A review of the literature showed cases of popliteal

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Figure 1. Maximum intensity projection (MIP) of the aorta and limb arteries. The first aneurysm was located to the infrarenal aorta and the second to the left common iliac artery and presented calcifications. The asymmetrical appearance of the femoral arteriomegaly is typical of KTS.

Figure 2. X-ray image of the lower limb extremity. Left cortical bone hypertrophy (1) and varicose veins (2) can be seen.

aneurysms, in particular those reported by Akagi et al.[3] This author reported also a unilateral arteriomegaly, but limited to the distal part of the superficial femoral artery. In our case, the histological analysis found, on the arterial wall, multiple calcified atherosclerotic and lipid plaques, fibrinocruoric thrombus and adventitial lymphocytic exocytosis. The different stains objectified deletion of the internal elastic lamina and diffuse mucoid soaking. Thus, there was an advanced state of 'destruction' of the arterial vasculature of the limb, as shown by the size of the aneurysms and also the diameter of the femoral vessels. The histological analysis was performed on tissue from the wall of the aneurysm, and did not detect any fragility of the media. That is the main difference with the case reported by Akagi et al.,[3] where the aneurysm wall showed intramural thrombus and disordered and disrupted elastic fibres in the extremely thinned media. Although in our case the histological signs are not specific of the KTS, the association of a unilateral iliofemoral arteriomegaly with venous and bones abnormalities of the same limb suggest a common pathogenesis. Moreover, there was another case of KTS with cerebral aneurysms, which reported also the presence of thrombus, calcifications and cholesterol deposits.[1] As KTS is caused by a mesodermal abnormality *in utero*,[4] the presence of venous and arterial aneurysms could be expected to exist. However, no other author reported such an advanced state of arterial wall destruction. It would have been interesting to obtain an analysis of the dilated wall on a non-aneurysmal area, to search for these anomalies. The other connective tissue disorders could not been excluded categorically because no immunohistochemical stains or gene testing were done; but, if the most recent diagnostic criteria[5] of KTS are considered, the bone hypertrophy associated with venous malformations is sufficiently typical to make the diagnosis of KTS. Arterial dilatation is an uncommon manifestation of KTS but extremely developed in the present case.

Ethical Approval

None.

Conflict of Interest/Funding

None.

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